# Disruption of the blood–perilymph barrier preceding endolymphatic hydrops formation in Meniere's disease

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Running heads : Disruption of the blood–perilymph barrier

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**Case presentation**

A 68-year-old woman was referred to our hospital with fluctuating sensorineural hearing loss (SNHL) in the left ear for 5 years and recurrent vertigo for 6 months. She also had unchanged SNHL at high frequencies in the right ear for 3 years. Middle ear pressure therapy (MEPT) was applied to her left ear using a novel transtympanic membrane massage device designed for treatment of Meniere's disease (MD) resistant to conservative treatment including vitamins, diuretics and intravenous or intratympanic steroids (1). Temporal change in endolymphatic hydrops (EH) and perilymphatic enhancement (PE) were evaluated during the course of treatment by magnetic resonance imaging (MRI) performed 4 hours after intravenous injection of gadolinium hydrate using a 3-T MRI scanner equipped with a receive-only, 32-channel, phased-array coil. Heavily T2-weighted three-dimensional fluid-attenuated inversion recovery MRI in 7 min was used to evaluate PE. HYDROPS (hybrid of reversed image of positive endolymph signal and native image of positive perilymph signal) was used to detect EH. (2) Semi-quantitative assessment of PE was performed by measurement of signal intensity ratios (SIRs) between values in the basal turn of the cochlea and those in the cerebellar hemisphere (3). High SIRs indicate disruption of the blood−perilymph barrier (BPB) in the cochlea as the cause of intractable inner ear disturbances (3), with reference to a recently proposed method in which the combination of EH and PE assessments showed high diagnostic accuracy for detecting MD (4).

MRI acquired prior to the middle ear pressure therapy revealed significant EH in the left cochlea and vestibule, but not on the right side (Fig. 1A). SIRs of PE in the right and left ears were 14.2 and 11.2, respectively (Fig. 1B). Based on left fluctuating SNHL, repeated vertigo symptoms and left EH on MRI, she was diagnosed with left MD. After 1 year of MEPT and reduction of medication, the number of episodes of vertigo per month decreased with stable hearing levels in the left ear, but hearing thresholds at lower frequencies (125,

250, and 500 Hz) fluctuated in both ears. MRI performed after 1 year revealed EH in the left ear; and SIRs of PE in the right and left ears were 12.1 and 9.5, respectively. Though fluctuating SNHL in the right ear suggested evolution to bilateral MD, vertigo symptoms were thought to be caused mainly by the left ear because no right EH was seen on MRI. After an additional 1 years of MEPT without medication, she was free of vertigo attacks and continued to have bilateral hearing fluctuations at low frequencies. On MRI performed 2 years after, SIRs of PE in the right and left ears had decreased to 9.9 and 7.5, respectively; however, development of EH was detected in the cochlea and vestibule of the right ear (Fig. 2A and 2B).

On the initial MRI when she had fluctuating SNHL only in the left ear, EH was observed only in the left ear, despite high SIRs of PE in the right ear too. SIRs of PE gradually decreased in both ears on the second and third MRI when she had fluctuating SNHL in the bilateral ear, but on the third MRI, EH formation was observed in the cochlea and the vestibule of the right ear. High PE would demonstrate alterations in the BPB with increased vascular permeability in ears with MD (3,5). Combination of EH and PE in the bilateral ear assessment has been proposed as a highly accurate method for the diagnosis of MD (4). In this study, High PE appeared prior to EH, suggesting that alterations of vascular permeability in the BPB may have led to the formation of EH.

## References

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**Figure legends**

Figure 1: Initial MRI shows SPACE (Sampling Perfection with Application optimized Contrast using different flip angle Evolution) sequence imaging (A), significant EH in the cochlea (white arrows) and vestibule (white arrowhead) of the left ear (B) and strong PE in the right and left cochlea (white arrows) (C).

Figure 2: MRI acquired 2 years later showed EH in the cochlea (white arrows) and vestibule (white arrowhead) of the right and left ears (A). The signal intensity ratios of PE in the right and left ears have decreased respectively (white arrows) (B).

Figure (TIFF, PDF, Word Doc, PPT, or EPS files are acceptable)

Figure1.tif

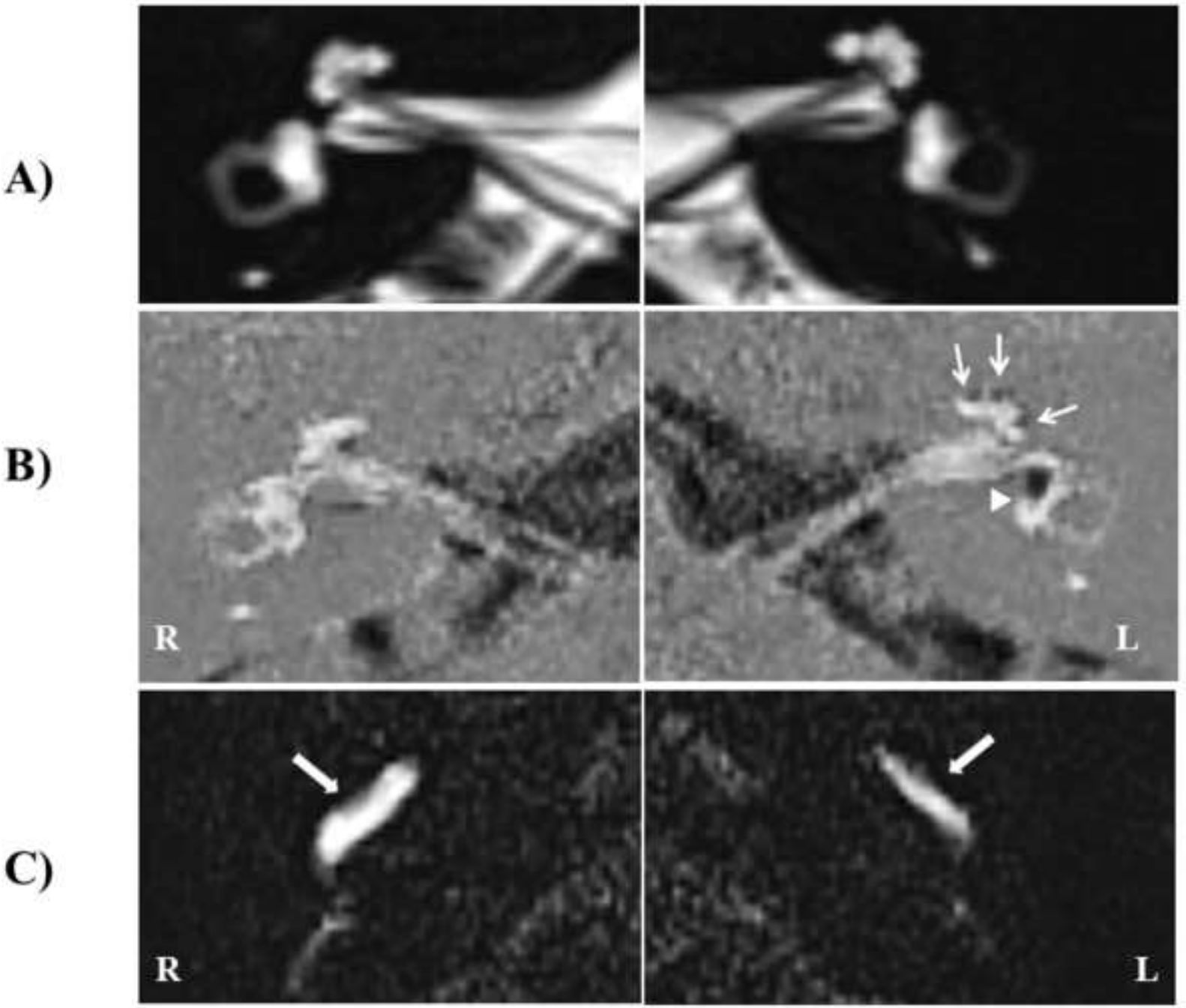


**Figure 1**

Figure (TIFF, PDF, Word Doc, PPT, or EPS files are acceptable)

Figure2.tif

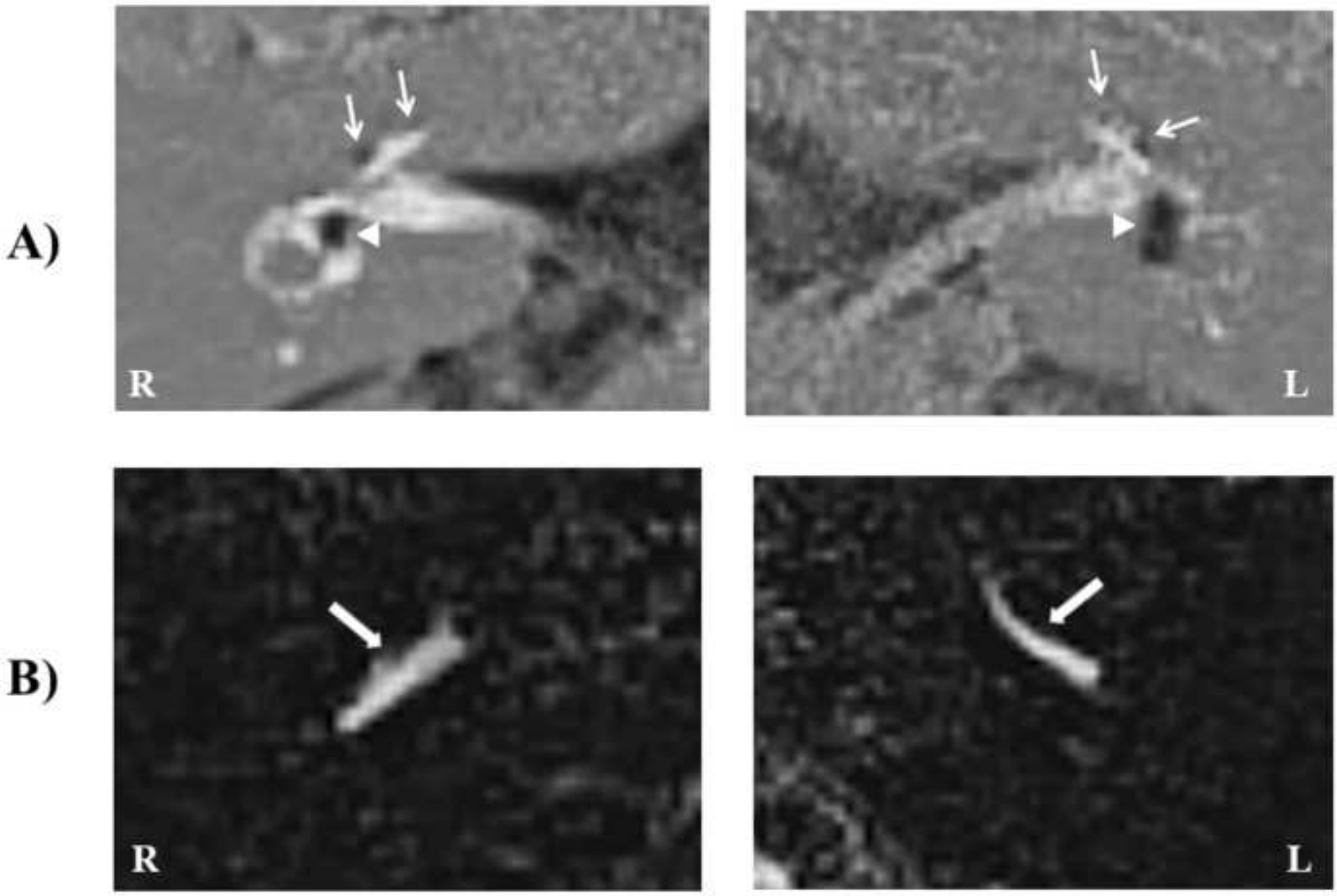


**Figure 2**